A unique case of a digital tourniquet in ichthyosis vulgaris

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Case Report

A unique case of a digital tourniquet in ichthyosis vulgaris

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ABSTRACT

We report a unique case of a digital tourniquet in a patient with ichthyosis vulgaris. We have identified no previous case reports documenting the occurrence of a digital tourniquet in patients caused by this condition. Ichthyosis vulgaris is a skin condition which causes increased scaling of the skin and in this case, resulted in the formation of a tourniquet-like circumferential constriction to one of the patient's digits.

Introduction

Ichthyosis vulgaris, the most prevalent subtype of ichthyosis, is a skin condition inherited in an autosomal dominant pattern with variable penetrance.[1] We report the first case of ichthyosis vulgaris causing a tourniquet-like constriction to a digit. Hair tourniquet of the digits is not an uncommon occurrence, particularly in children, however, the management of this case required a very different approach to that of a simple external constricting body.

Case report

A 41 year old female patient presented to our department with a tourniquet-like contracture surrounding her right little finger caused by ichthyosis vulgaris. A band of skin causing progressing contracture at the base of the right little finger had been developing over several months. The patient

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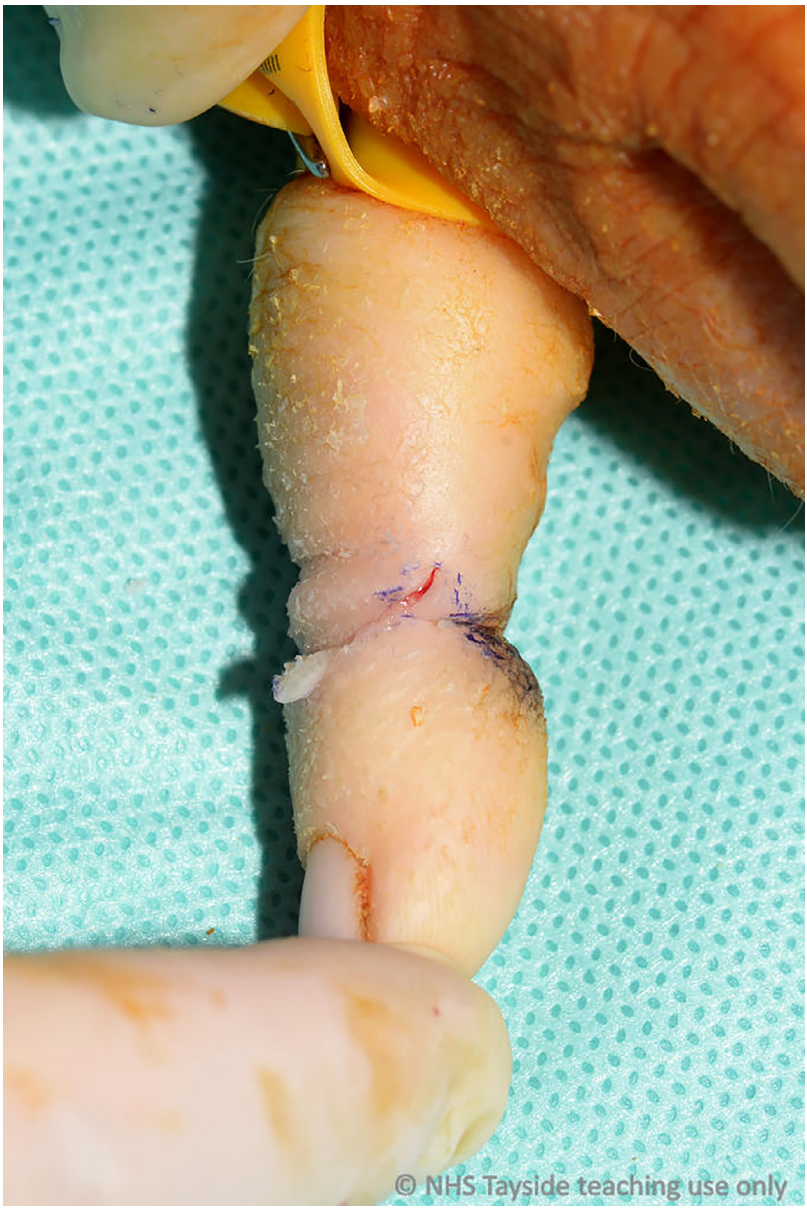
Figures 1–6. Intra-operative photographs displaying a constricting band to the digit caused by Ichthyosis Vulgaris. The constricting band was surgically released and the skin defect reconstructed with a full thickness skin graft.

presented with acute tightening of the contracture at the base of the finger with paraesthesia and potentially compromised perfusion to the digit.

The patient reported lifelong symptoms associated with ichthyosis vulgaris and her other past medical independently history included hidradenitis superitiva.

After informed consent the patient was taken to theatre as an emergency procedure and underwent release of the contracting band of skin (see Figures 1–6). This necessitated reconstruction in the



Figures 1–6. Continued

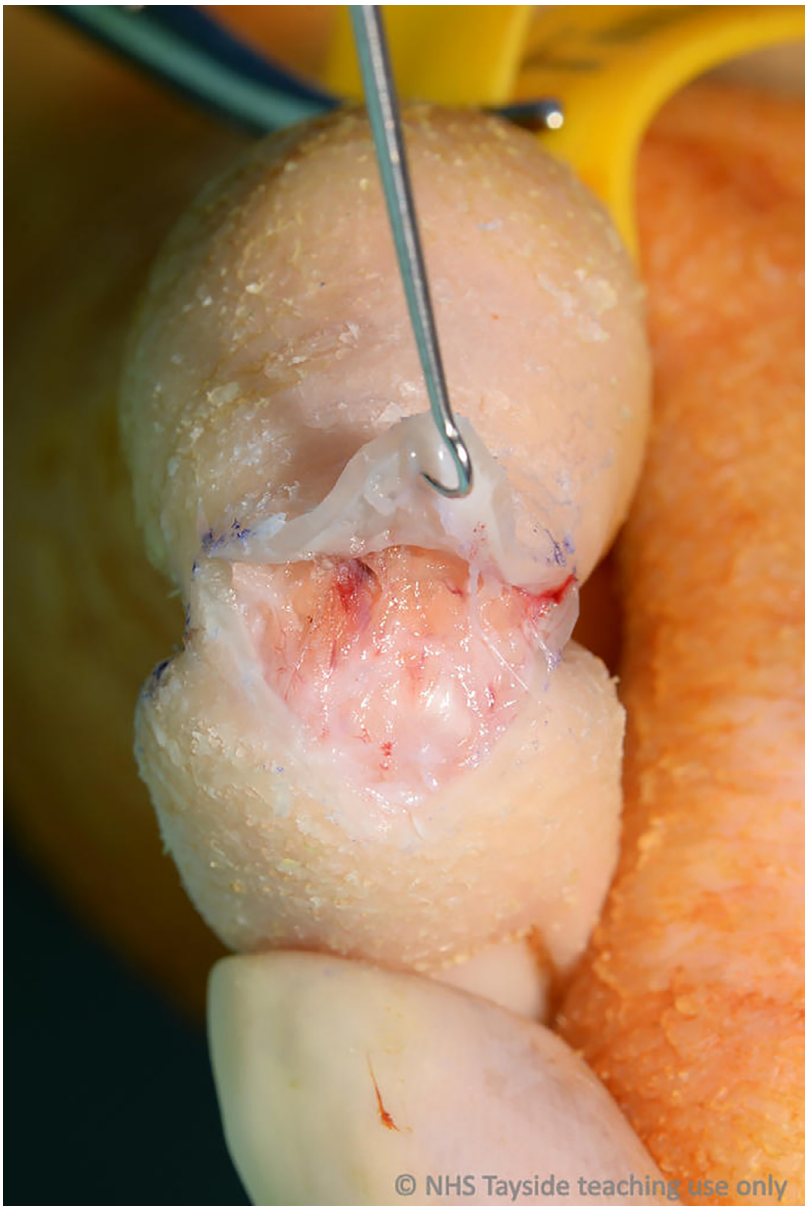
form of a full thickness skin graft, harvested from the groin. The patient had no history of hidradenitis superitiva disease in the groin.

The patient was followed up at six months and had no recurrence of constriction at the operated area (see Figures 7–12). There was full range of movement in this finger with no pain and no paraesthesia in the digit. However, the patient had developed a constricting band in another digit of the same hand and was listed for release of this.

Figures 1–6. Continued

Discussion

The condition has an estimated prevalence of 4.0–7.7% in European populations.[2] It is thought that 1 in 250–1000 people have loss of function mutations of the FLG gene which may predispose them to developing some form of ichthyosis.[1] It causes disorder of keratinisation which preferentially affects the palmar and plantar tissue.[3]



Figures 1–6. Continued

The protein filaggrin enables the terminal differentiation of the epidermis and thus formation of an adequate skin barrier.[4] This differentiation of the human epidermis is reliant on a series of genes on chromosome 1q21, known as the epidermal differentiation complex (EDC).[5] Loss of function mutations of the filaggrin gene (*FLG*) result in xerosis, keratosis pilaris, scaling and hyperlinearity.[6] Cells accumulate in the stratum corneum creating scaling of the skin.[4] The condition manifests itself clinically as excessively dry and scaly skin.[2]

Figures 1–6. Continued

Figures 1–6. Continued

Ichthyosis vulgaris tends to develop in the months following birth.[7] The condition is more common in Northern climates and in fair coloured skin and there are reports the condition may improve in warmer weather conditions.[1,6] Ichthyosis vulgaris has been strongly linked to atopic disorders and affected individuals are particularly prone to developing dermatitis.[8,9] Genetic testing may be used for confirmation of the diagnosis and subsequent counselling.[1]

Histological analysis of skin biopsies of ichthyosis shows a reduction or absence of the stratum granulum, reduced keratohyalin granules and orthokeratosis[7,10]

Medical management is the mainstay of treatment for ichthyosis vulgaris, focusing on minimising skin scaling, improving skin hydration and promoting skin barrier function [I]. This is achieved

Figures 7–12. Post-operative photographs displaying a well healed full thickness skin graft and resolution of the tight constricting band.

primarily with moisturisers, emollients, topical steroids and keratolytic agents.[1,11] Reduction of environmental or contact irritants is also advocated.[11]

We report the first case of ichthyosis vulgaris causing a tourniquet-like constriction to a digit. Hair tourniquet of the digits is not an uncommon occurrence, particularly in children, however, the management of this case required a very different approach to that of a simple external constricting body. In order to decompress the tight skin band and reconstruct the base of the digit, the use of autologous

Figures 7–12. Continued

tissue was required, in this case a full thickness skin graft. The use of this graft and the chosen site was selected carefully, given this patients other comorbid condition, hidradenitis superitiva. Additionally, the patient was counselled about the rarity of her presentation and the limited knowledge of the outcomes following this procedure. Release of this constricting band does not address the underlying ichthyosis vulgaris and therefore it is impossible to predict the long term outcomes of full thickness grafting or indeed predict the patient's risk of further constricting bands in other digits.

Figures 7–12. Continued

Declaration of patient consent

The authors confirm that they have sought consent from the patient to use their images and clinical information to be reported. The patient understands that their name and initials will not be published.

Figures 7–12. Continued

Figures 7–12. Continued

Figures 7–12. Continued

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None.

Declaration of Competing Interest

There are no conflicts of interest.

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